all pigs. Myocardial perfusion in the normally-perfused LAD and collateral-perfused LCx regions were determined with colored microspheres. Coronary microvessels were studied *in vitro* with video-microscopy. Responses = % relaxation of acetylcholine-induced contraction.

Group	ADP (10 μM)	Pinacidil (10 μM)	SNP (10 μM)
LAD-control	78 ± 3*	83 ± 1	73 ± 4
LCx-control	46 ± 4	82 ± 4	76 ± 3
LAD-VEGF	77 ± 7*	84 ± 4	82 ± 6
LCx-VEGF	75 ± 3*	92 ± 4	78 ± 5

*p < 0.05 vs LCx-control.

Relaxations to pinacidil (K^+ ATP channel opener) and nitroprusside (SNP) were similar in all groups. Endothelium-dependent relaxation to ADP was impaired by collateral-dependent perfusion. These responses were significantly preserved with VEGF treatment. LCx/LAD blood flow ratio at rest was similar in the VEGF (1.10) and control (1.15) groups, but during atrial pacing, perfusion was improved in the VEGF-treated animals (1.50) compared with control pigs (0.98, $p < 0.01$ vs VEGF). The improved vascular function and perfusion associated with VEGF treatment may have implications regarding the management of patients with severe coronary disease who are not candidates for conventional methods of revascularization.

909 Tetralogy of Fallot: Etiology, Management, and Follow-Up

Monday, March 25, 1996, Noon–2:00 p.m.
Orange County Convention Center, Hall E
Presentation Hour: Noon–1:00 p.m.

909-22 Tetralogy of Fallot With and Without Pulmonary Atresia: Different Prevalence of Genetic Syndromes

Maria C. Digilio, Bruno Marino, Sabina Grazioli, Rita Mingarelli, Giuseppe Novelli, Aldo Giannotti, Bruno Dallapiccola. *Cardiology and Genetics, Bambino Gesù Hospital, Rome, and Tor Vergata University, Rome, Italy*

Tetralogy of Fallot (TF) with or without pulmonary atresia (PA) is a conotruncal heart defect occurring as isolated malformation or in association with noncardiac anomalies. In order to evaluate the prevalence of genetic syndromes, 168 consecutive children with TF underwent phenotypic, cardiological and genetic evaluation. Southern hybridization with HD7k probe and fluorescent in situ hybridization with Sc11.1 probe for detection of 22q11 deletion (Del22) were performed. Clinical and molecular results obtained in 22 patients with TF and PA have been compared with those found in 146 patients with classic TF. The prevalence of genetic syndromes among patients with TF and PA (14/22 = 64%) was significantly higher ($p < 0.01$) in comparison with that found in patients with classic TF (45/146 = 31%). A clinical diagnosis of DiGeorge, velocardiofacial or CHARGE syndromes was reached in 12/22 (54%) of patients with TF and PA and in 17/146 (12%) with TF ($p < 0.001$). Also the detection of Del22 was significantly different ($p < 0.01$) in the two groups (7/22 = 32% versus 9/146 = 6%). In conclusion, patients with TF and PA are at particular risk for genetic syndromes, especially branchial arch syndromes related to Del22. An accurate phenotypic and genetic evaluation is needed in these patients.

909-23 Isolated Conotruncal Heart Defects Are Really Related to Microdeletion of Chromosome 22q11?

Maria C. Digilio, Bruno Marino, Rita Mingarelli, Giuseppe Novelli, Francesca Amati, Aldo Mari, Aldo Giannotti, Bruno Dallapiccola. *Cardiology and Genetics, Bambino Gesù Hospital, Rome, and Dept. Cell. Biology, Tor Vergata University, Rome, Italy*

Cytogenetic and molecular studies have documented a strong association between deletion of chromosome 22q11 (Del22) and conotruncal heart defects (CTHDs) in the setting of DiGeorge (DG) and Velocardiofacial (VCF) syndromes. Del22 has been reported also in a high proportion (15–30%) of children with isolated non-syndromic CTHD. In order to investigate the effective prevalence of Del22, we screened 183 patients with strictly isolated CTHD. Children with classic or subtle phenotypic anomalies, abnormal facies, thymic defects, cleft palate and hypocalcemia were excluded from this study. Tetralogy of Fallot was present in 100 patients, transposition (TGA) in 42, congenitally corrected TGA in 14, pulmonary atresia with ventricular septal defect in 10, double outlet right ventricle in 6, asplenia in 6, truncus in 5. Testing included Southern hybridization with probe HD7k and in situ hybridization (FISH) with probe Sc11.1. Del22 was detected in only one patient with TGA (1/183 = 0.5%). In contrast with previous reports, the prevalence of Del22 is very low in non-syndromic children with strictly isolated CTHD. An accurate phenotypic analysis and clinical follow up may select the patients with CTHD at substantial risk for Del22.

909-24 Determinants of Long-Term Survival in Tetralogy of Fallot: 36 Years Follow-Up in 447 First Year Survivors After Repair

Georg Nollert, Teddy Fischlein, Eckart Kreuzer, Armin Welz, Heinrich Netz, Werner Klinner. *Bruno Reichart University of Munich, Germany*

We investigated ultra-long-term survival after surgical repair of tetralogy of fallot (TOF) in order to analyze which parameters predispose or cause for premature death. From 12/1958 to 6/1977 701 patients (age: 11.2 ± 9.2 years; range 0–56 years; 44% female) had a correction of their TOF at our institution; 14% (n = 116) were lost to follow-up. One previous palliative operation had 33.6% of the patients; 2.6% had two and 0.1% three. In the operative period 124 patients (25%) died in the first year after surgical repair. These patients were excluded for further statistical analysis. Actuarial 10, 20, 30 and 36 years survival was 97%, 94%, 91% and 91% respectively. Univariate correlates of survival in the remaining 447 patients were use of a pulmonary outflow patch (n = 95; p = 0.0004), cardio-pulmonary bypass time (operations using ventricular fibrillation; $p < 0.0001$), age ($p < 0.0001$; lowest risk between 6 and 20 years) and date of operation (better outcome of patients operated in the 1970ies; p = 0.026); multivariate correlates in a Cox regression model were only bypass time (p = 0.035) and use of a pulmonary outflow patch (p = 0.030). Patients with a bypass time below 40 min and no pulmonary outflow patch (n = 128) had a 36 years actuarial survival of 96% with only 1 death at 25 years after the operation and reached normal life expectancy. Ischemic damage during the operation in these chronically hypoxic hearts (average age in this historical group above 10 years) and pulmonary insufficiency due to an outflow patch are the most important factors influencing the excellent long-term outcome after surgical repair of TOF.

909-25 Long Term Benefit of Pulmonary Valve Replacement for Progressive Pulmonary Insufficiency After Repair of Tetralogy of Fallot

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Pulmonary valve replacement (PVR) is frequently considered for pts with progressive pulmonary insufficiency (PI) and severe right ventricular (RV) dilation after repair of Tetralogy of Fallot. Short term benefit has been documented but long term results remain unclear especially in light of reports of xenografts requiring re-replacement for valve degeneration and/or stenosis. To evaluate the long term response to PVR in this setting, we reviewed the results of a group of 12 pts who have been followed prospectively for now a mean of 10 yrs after PVR. Mean age at Tetralogy repair was 6.6 yrs (range: 3.5 to 10 yrs). In each pt, the decision to proceed to PVR was based on progressive RV dilation on echocardiography, plus new onset of tricuspid insufficiency in 3 of 12 and stenosis and insufficiency of a previously placed conduit in [illegible]. Mean age at PVR was 14.3 yrs (range: 8.5 to 21 yrs). Preoperative, 2 [illegible] 10 yr post-operative findings on echocardiography, chest x-ray, and Holter monitoring are shown below:

	Pre-op (n = 12)	2 yrs post (n = 12)	10 yrs post (n = 10)#
Echo RV/LV	1.02 ± 0.25	0.74 ± 0.14*	0.79 ± 0.17*
C/T Ratio	0.59 ± 0.02	0.55 ± 0.02*	0.54 ± 0.05*
Holter:Gr 4 VPCs	3/12	3/12	3/10

*p < 0.01 compared with pre-op.
#Two pts demonstrated progressive stenosis and insufficiency of the conduit valves at 5 yrs after PVR and had a second PVR performed with again reduction in RV size. At 10 yrs after their original PVR, the echo, x-ray and holter findings in these two pts do not differ from those of the group at large.

These study findings indicate objective improvement in RV size late after PVR for progressive PI post repair of Tetralogy of Fallot.

909-26 Pulmonary Artery Stenosis in Infant Repairs of Tetralogy of Fallot

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Controversy continues over the repair of Tetralogy of Fallot (TOF) in the first year of life where undersized pulmonary arteries (PA) may lead to higher rate of transannular patching and subsequent PA stenoses. A lower mortality rate